

Immunotag™ PRIO Polyclonal Antibody

Antibody Specification	
Catalog No.	ITN2009
Product Description	Immunotag™ PRIO Polyclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	PRIO
Clonality	Polyclonal
Storage/Stability	-20°C/1 year
Application	WB,ELISA
Recommended Dilution	WB 1:500-2000 ELISA 1:5000-20000
Concentration	1 mg/ml
Reactive Species	Human,Rat,Mouse
Host Species	Rabbit
Immunogen	Synthesized peptide derived from part region of human protein
Specificity	PRIO Polyclonal Antibody detects endogenous levels of protein.
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen
Form	Liquid in PBS containing 50% glycerol, and 0.02% sodium azide.
Gene Name	PRNP PRIP PRP
Accession No.	P04156 P04925 P13852

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Description	prion protein(PRNP) Homo sapiens The protein encoded by this gene is a membrane glycosylphosphatidylinositol-anchored glycoprotein that tends to aggregate into rod-like structures. The encoded protein contains a highly unstable region of five tandem octapeptide repeats. This gene is found on chromosome 20, approximately 20 kbp upstream of a gene which encodes a biochemically and structurally similar protein to the one encoded by this gene. Mutations in the repeat region as well as elsewhere in this gene have been associated with Creutzfeldt-Jakob disease, fatal familial insomnia, Gerstmann-Straussler disease, Huntington disease-like 1, and kuru. An overlapping open reading frame has been found for this gene that encodes a smaller, structurally unrelated protein, AltPrp. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Nov 2014],
Cell Pathway/ Category	Prion diseases,
Protein Expression	Blood,Brain,Ovary,Prostate,
Subcellular Localization	nucleus,cytoplasm,mitochondrion,mitochondrial outer membrane,endoplasmic reticulum,Golgi apparatus,plasma membrane,cell surface,integral component of membrane,extrinsic component of membrane,anchored component of membrane,membrane raft,

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Protein Function	disease:Defects in PRNP are the cause of Creutzfeldt-Jakob disease (CJD) [MIM:123400]. CJD occurs primarily as a sporadic disorder (1 per million), while 10-15% are familial. Accidental transmission of CJD to humans appears to be iatrogenic (contaminated human growth hormone (HGH), corneal transplantation, electroencephalographic electrode implantation, etc.). Epidemiologic studies have failed to implicate the ingestion of infected animal meat in the pathogenesis of CJD in human. The triad of microscopic features that characterize the prion diseases consists of (1) spongiform degeneration of neurons, (2) severe astrocytic gliosis that often appears to be out of proportion to the degree of nerve cell loss, and (3) amyloid plaque formation. CJD is characterized by progressive dementia and myoclonic seizures, affecting adults in mid-life. Some patients present sleep disorders, abnormalities of high cortical function, cerebellar and corticospinal disturbances. The disease ends in death after a 3-12 months illness.,disease:Defects in PRNP are the cause of fatal familial insomnia (FFI) [MIM:600072]. FFI is an autosomal dominant disorder and is characterized by neuronal degeneration limited to selected thalamic nuclei and progressive insomnia.,disease:Defects in PRNP are the cause of Gerstmann-Straussler disease (GSD) [MIM:137440]. GSD is a heterogeneous disorder and was defined as a spinocerebellar ataxia with dementia and plaquelike deposits. GSD incidence is less than 2 per 100 million live births.,disease:Defects in PRNP are the cause of Huntington disease-like 1 (HDL1) [MIM:603218]. HDL1 is an autosomal dominant, early onset neurodegenerative disorder with prominent psychiatric features.,disease:Defects in PRNP are the cause of kuru [MIM:245300]. Kuru is transmitted during ritualistic cannibalism, among natives of the New Guinea highlands. Patients exhibit various movement disorders like cerebellar abnormalities, rigidity of the limbs, and clonus. Emotional lability is present, and dementia is conspicuously absent. Death usually occurs from 3 to 12 month after onset.,disease:Defects in PRNP are the cause of prion disease with protracted course [MIM:606688]; an autosomal dominant presenile dementia with a rapidly progressive and protracted clinical course. The dementia was characterized clinically by frontotemporal features, including early personality changes. Some patients had memory loss, several showed aggressiveness, hyperorality and verbal stereotypy, others had parkinsonian symptoms.,disease:PrP is found in high quantity in the brain of humans and animals infected with neurodegenerative diseases known as transmissible spongiform encephalopathies or prion diseases, like: Creutzfeldt-Jakob disease (CJD), fatal familial insomnia (FFI), Gerstmann-Straussler disease (GSD), Huntington disease-like 1 (HDL1) and kuru in humans; scrapie in sheep and goat; bovine spongiform encephalopathy (BSE) in cattle; transmissible mink encephalopathy (TME); chronic wasting disease (CWD) of mule deer and elk; feline spongiform encephalopathy (FSE) in cats and exotic ungulate encephalopathy (EUE) in nyala and greater kudu. The prion diseases illustrate three manifestations of CNS degeneration: (1) infectious (2) sporadic and (3) dominantly inherited forms. TME, CWD, BSE, FSE, EUE are all thought to occur after consumption of prion-infected foodstuffs.,function:The physiological function of PrP is not known.,online information:PRNP entry,polymorphism:The five tandem octapeptide repeats region is highly unstable. Insertions or deletions of octapeptide repeat units are associated to prion disease.,PTM:The glycosylation pattern (the amount of mono-, di- and non-glycosylated forms or glycoforms) seems to differ in normal and CJD prion.,similarity:Belongs to the prion family.,subunit:PrP has a tendency to aggregate yielding polymers called "rods". Interacts with GRB2, PRNIP and SYN1.,
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